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Letter to the Editor

The value of performing invasive risk stratification in young patients with the Brugada syndrome

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The Brugada syndrome is an inherited disease characterised by coved-type ST-segment elevation in the right precordial leads and increased risk for sudden cardiac death.[1] The phenotypic expression of the disease extends from the completely asymptomatic state to potentially lethal arrhythmias and sudden cardiac death. The disease typically manifests in the fourth decade of life, but severe cases have shown clinical expression during childhood and can result in life-threatening arrhythmias.[2,3]

A provocative test consisting of intravenous administration of a class IC antiarrhythmic drug is a well-established tool to unmask the diagnosis of the type I pattern in young patients with suspected Brugada syndrome and a non-diagnostic electrocardiogram.

In contrast, risk stratification by means of ventricular stimulation in this population remains controversial.

The prevalence of the Brugada syndrome is extremely low in the paediatric population (0.009%)[4]; moreover, symptoms present in only 30% of young patients with the Brugada syndrome. Although rarely diagnosed in children, the Brugada syndrome can manifest as sudden cardiac death at a very young age.[5]

There is thus a critical need to accurately identify young patients at increased risk for cardiac arrest, both to minimise mortality and to maintain a low morbidity related to unnecessary device implantation.

Andorin et al recently reported on a multicentric European study that included 106 patients aged <19 years.[6] A baseline electrophysiological study was performed in 22 (21%) patients on the clinical advice of an expert cardiologist, with a positive result in nine (41%).

Our group has recently reported on the feasibility and outcomes of ventricular stimulation in young patients.[3] In this series including a cohort of 128 patients <25 years, 28 symptomatic patients underwent a ventricular stimulation protocol, and sustained ventricular fibrillation, or polymorphic ventricular tachycardia was induced in 6 (21%). No ventricular arrhythmias were induced in the 70 ventricular stimulation protocols performed in asymptomatic patients. No complications resulted from the cardiac catheterisation or from the arrhythmia induction.

Performing a single-catheter cardiac catheterisation under propofol sedation has been proven to be a safe procedure when performed by experienced hands and in the appropriate institutional setting, with the provision of advanced life-support facilities. The protocol should include a complete electrophysiological study that provides valuable data on the general electrical performance of the heart. The study should start by measuring the baseline intervals, including the AH and HV intervals. The sinus node function should also be evaluated in all patients, by means of sinus node recovery time. The atrioventricular conduction system can be evaluated by measuring the Wenkebach cycle length and the atrioventricular nodal effective refractory period. In patients with either a history of palpitations or with evidence of supraventricular tachycardia from Holter monitoring, an atrial stimulation protocol should be performed. The study concludes with a ventricular stimulation protocol consisting of a maximum of three ventricular extra-stimuli, with a minimum coupling interval of 200 ms, delivered from a single right ventricular site. Results are considered positive in the case of induction of sustained ventricular arrhythmias. A ventricular effective refractory period <200 ms and HV interval >60 ms are highly suggestive of the Brugada syndrome in adults, but need further investigation in the younger age group.

In the case of a negative result, the presence of an abnormal electrical impulse generation or propagation provides invaluable confirmation of the clinical expression of the disease and helps guide the follow-up.

Because of scarce reports on the Brugada syndrome in the paediatric age, it is not possible to accurately define the value of programmed ventricular stimulation for risk stratification. No systematic studies have yet been performed to understand the mid- and

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long-term risks of a positive ventricular stimulation in young patients; further, the ideal age to start performing invasive risk stratification is unknown.

As shown by the study by Andorin et al,[7] different centres seem to be using different protocols.

It has been our institutional practice to recommend risk stratification by means of ventricular stimulation in young patients with severe syncope and a diagnosis of the Brugada syndrome by means of spontaneous or drug-induced type I electrocardiography. In this age group, a patient-oriented management approach should be considered on an individual basis, taking into consideration the clinical circumstances and preferences of the family. We believe that by constructing a large and standardised database, we are building evidence that in the long term will help better understand this enigmatic disease and guide therapeutic decisions.

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